

Trabecular variant of juvenile aggressive ossifying fibroma

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Abstract

Juvenile aggressive ossifying fibroma (JAOF) is an uncommon benign lesion which is distinctly aggressive in behavior with high tendency for recurrence. It appears in early age and in 79% of patients is diagnosed before 15. It has two histological variants: psammomatoid and trabecular, with the latter being less common with a stronger tendency to recur. In this article, we present a case of trabecular JAOF, in which treatment could not be given despite the availability of all requisites for surgery and good financial status, due to parental indifference and negligence.

Case Report

A 8-year-old male patient reported to the outpatient department with the main complaint of swelling of left side of face. The patient's parents gave a history of a rapidly growing painless swelling, which first became apparent around a month before. There was no history of any trauma or tooth pain. On general examination, mild pallor and weight loss were the only apparent findings. Facial asymmetry was distinguishable with an expansile lesion of about 4×5 cm in size, involving left side of face. The anterior extent was along a line drawn perpendicular to medial canthus of left eye. Posteriorly, the extent was up to the anterior border of masseter muscle. Superior and inferior boundaries corresponded to infra-orbital margin as well as zygomatic arch and a line joining left corner of oral commissure with the point of attachment of ear lobule, respectively. The overlying skin was normal. The left ala of nose was elevated with slight reduced display of vermillion of lateral half of upper lip. Bilateral nasal patency was present. Palpation revealed an underlying non-tender, bony hard swelling with smooth texture. Paresthesia was absent with no associated lymph node involvement. Intra-orally, a spherical swelling was apparent, obliterating left maxillary vestibule and extending from lateral incisor to deciduous second molar (Figure 1A). The overlying mucosa was normal in appear-

ance. None of the teeth were mobile, carious or non-vital.

Investigations advised were routine blood picture, serum alkaline phosphatase determination and computed tomography (CT). An orthopantomograph was also ordered to evaluate the status of dentition. The biochemical profile came to be within normal range. Sections of CT showed a destructive expansile lesion involving left maxilla with well delineated cortical boundary. No involvement of maxillary sinus was evident on (Figure 1B).

Incisional biopsy was done and adequate sample was obtained from the lesion and sent for histopathological examination. Microscopic examination showed large area of cellular fibrous tissue with plump, ovoid to spindle fibroblasts and some areas of collagen. Irregular islands of immature bony tissue along with mature bony trabeculae were also seen embedded in stroma along with scattered multinucleated giant cells (Figure 1C). Based on this picture, supplemented by the clinical presentation and radiographic findings, a diagnosis of JAOF of trabecular variety was made.

Keeping in mind the extent of the lesion, it was decided to enucleate the lesion with simultaneous peripheral osteotomy through Weber-Ferguson incision. Along with the treatment plan, the patient's parents were explained the aggressive nature of the lesion and its clinical outcomes if left untreated and asked to sign the consent form. To our surprise, the parents declined the treatment proposed. The reason they cited was the resulting scar from the incision and complexity of the operative procedure requiring large amount of bone removal. The parents were unsuccessfully persuaded to get the treatment done. Eventually we had to let go of the patient without any treatment.

Discussion

Juvenile aggressive ossifying fibroma is an uncommon benign lesion which is distinctly aggressive in behavior with a high tendency for recurrence and is usually seen affecting younger individuals with age less than 15 years.¹⁻³ Ninety percent of the lesions located in the face region, involve the sinuses, mainly the maxillary antra.¹ It can be distinguished from standard ossifying fibroma based on its clinically aggressive biologic behavior and occurrence in younger individuals and also by less frequent involvement of mandible. The lesion presents with proptosis, exophthalmos, visual disturbances, nasal obstruction and facial asymmetry. Radiographic features are non-specific and depending on the location of the tumor, maturation stage and stage of

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ossification, they are uni- or multilocular well-defined lesions which may be radiolucent, mixed or radiopaque.⁴ Aggressive lesions may show cortical thinning and perforation.⁵ Psammomatoid JOF is reported more commonly than trabecular JOF.^{6,7} Psammomatoid JOF occurs predominantly in the sino-nasal and orbital bones, and trabecular JOF predominantly affects the jaws. Psammomatoid JOF has aggressive behavior and it has a very strong tendency to recur.^{8,9} Johnson *et al.*, in a review of 3000 fibro-osseous lesions, found that a majority of tumors were located in facial bones, among which approximately 90% originated from paranasal sinuses.¹⁰ When jaws are involved, the maxilla precedes the mandible in frequency, as seen in the present case. Trabecular JOF is distinguished by the presence of trabeculae of fibrillar osteoid and woven bone and psammomatoid JOF is characterized by the presence of small uniform spherical ossicles that resemble psammoma bodies.⁸ The reported case in this article presented with microscopic features consistent with the trabecular type and hence the diagnosis.

The differential diagnosis should include fibrous dysplasia, Burkitt lymphoma, osteosarcoma and cystic lesions. Fibrous dysplasia was ruled out by the rapid growth, monostotic involvement, and the well-delineated radiographic margins. To avoid misdiagnosis and mismanagement, it is prudent to differentiate it from Burkitt lymphoma which presents with gross mobility and displacement of the associated teeth in addition to varying abdominal symptoms, which may range from

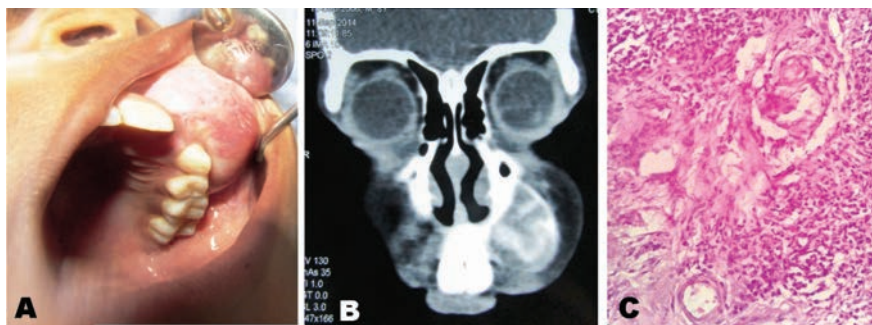


Figure 1. A) Intraoral view of the lesion demonstrating the extent of the lesion. B) Coronal section of computed tomography scan demonstrating the expansile, destructive lesion with cortical boundary. C) Histo-pathological picture of the lesion demonstrating large area of cellular fibrous tissue with plump, ovoid to spindled fibroblasts and some areas of collagen and irregular islands of immature bony tissue along with mature bony trabeculae embedded in stroma along with scattered multinucleated giant cells.

splenomegaly, hepatomegaly or both. The present case shared few clinical similarities with Burkitt lymphoma however, other distinct clinical features of Burkitt lymphoma were absent. Osteosarcoma was ruled out by benign radiographic appearance, absence of cytological atypia, and infiltration of adjacent structures.

Management of JAOF requires complete surgical excision taking into consideration, the size, location and extent of the tumor. Small, accessible lesions may be amenable to surgical excision alone or with peripheral ostectomy. However, larger or recurrent lesions necessitate a more aggressive procedure such as en bloc resection which may create considerable deformity especially if the tumor has invaded the orbit or the cranial cavity. In the present case, we decided to surgically excise the lesion along with peripheral ostectomy to prevent recurrence through Weber-Ferguson approach. Despite the availability of all the requisites for the surgery and good financial status of the patient's family, the treatment had to be deferred as the patient's parents did not give

their consent for the surgery when approached for the same as the patient was a minor. The parents were clearly explained the treatment plan as well as the clinical outcome of the disease if left untreated but they disagreed citing the unaesthetic scar and operative procedure requiring removal of considerably large amount of their child's facial bone as the main reason for their un-approval. Another reason they gave was that the patient was their only child and they did not want to put him under the risk of a major operative procedure.

It is not unusual for parents to become extra protective towards their only child especially when dealing with a potentially threatening disease. We believe that surgical counseling should be given not only by the consulting team but a qualified psychiatrist also should be a part of the counseling team as very often the surgeons fail to see the psychological impact on the patient and his/her family which a psychiatrist won't oversee.

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